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CONTENTS

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## ORIGINAL ARTICLES

- |   |   |
|---|---|
| Neoplasms involving the spinal cord in Zimbabweans: an analysis of 262 cases .....  | PI Garrido, S Laher-Mooncey, NL Murphree, N Jonker, LF Levy, S Makarawo ..... 201 |
| Termination of pregnancy using extra-amniotic prostaglandin F <sub>2α</sub> - experience in a peripheral hospital .....                                 | A Krolkowski, KA Janowski, JV Larsen ..... 204                                    |
| Epidemiological study of drug use in urban and rural secondary schools in Zimbabwe .....  | SW Acuda, AH Eide ..... 207   |
| A comparison of three types of "M" traps for sampling tsetse fly (Diptera: Glossinidae) populations at South Luangwa Game Management Area, Zambia ..... | MM Ziba, M Mukunyandela, S Siziya, JS Chipipa, L Mwanza ..... 212                 |
| Increased susceptibility of erythrocyte membrane lipids to peroxidation in sickle cell disease .....  | EU Essien ..... 217   |

## CASE REPORTS

- |   |  |
|---|--|
| Intracranial Burkitt's lymphoma with extension into orbital spaces resulting in bilateral blindness | K Ramesh, LD Gahukambele, DB Gahukambele ... 220 |
| Munchausen syndrome by proxy - a case report and review of literature .....                         | OA Oyelami, AM Alhaj, IK Airede ..... 222        |
| Hypothyroxinemia in Acquired Immune Deficiency Syndrome (AIDS) .....                                | CC Ezcala, E Chukwurah ..... 226                 |

## REVIEW ARTICLES

- |  |                                      |
|--|--------------------------------------|
| Advances in electrodiagnosis in Zimbabwe. Part II: "Evoked Potentials" ..... | O Parry, LF Levy, J Mielke ..... 229 |
|--|--------------------------------------|

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CASE REPORTS

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## **Intracranial Burkitt's lymphoma with extension into orbital spaces resulting in bilateral blindness.**

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### **SUMMARY**

We report a rare case of primary Burkitt's lymphoma involving the supra and parasellar regions with extension of the tumour into frontal lobes and posterior orbital spaces resulting in bilateral proptosis and blindness in a four year old male child.

### **INTRODUCTION**

Burkitt's lymphoma commonly affects the jaw and the abdominal organs. It is well known that Burkitt's lymphoma affects the central nervous system during the relapse,<sup>1,3</sup> yet its occurrence as a primary intracranial tumour without the involvement of other organs is extremely rare.<sup>4-6</sup> In this report, we present a four year old child who presented with a grotesque appearance due to bilateral proptosis, and total blindness.

### **CASE REPORT**

A four year old male child was brought to the hospital with a history of blurring followed by loss of vision in both the eyes, which occurred over two and half months. The patient had a history of headache for the preceding year. Protrusion of both the eyes developed quite rapidly over the week prior to admission. There was no history of injury or epilepsy.

The child was emaciated with bilateral proptosis and periorbital oedema. The fundal examination revealed bilateral optic nerve atrophy. There was no other cranial neuropathies or other neurological signs. Exami-

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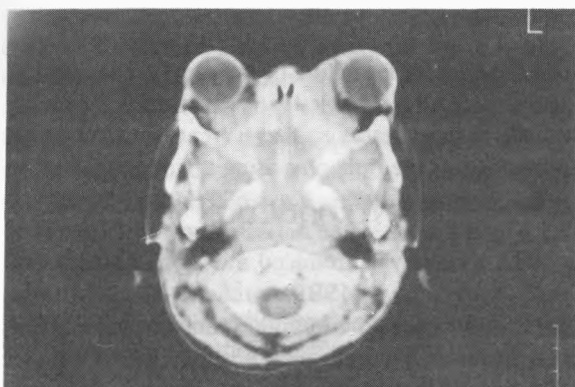
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nation of the jaw and abdomen showed no swelling. There was no peripheral lymphadenopathy.

Routine biochemical and haematological investigations including bone marrow aspiration were within normal limits. The serology was negative for human immuno-deficiency virus. Antibody titres for Epstein-Barr virus were not done due to lack of facilities. The cerebrospinal fluid examination revealed occasional abnormal lymphoid cells but no definite evidence of malignant pleocytosis. Chest X-ray was normal. Ultrasonography and computed tomography (CT) scan of the abdomen showed no evidence of tumour.

CT scanning of the brain with contrast was also done which showed an extensive homogenous hyperdense area with an irregular outline occupying the supra and parasellar regions measuring about 7.5 x 5 x 3 cms with extension into the frontal lobes, nasal, sphenoidal and epipharyngeal spaces. The mass was seen extending into both the posterior orbital spaces (Figure I). The anterior clinoid process was destroyed and the basal cistern was compressed anteriorly.

*Figure I: CT scan of brain showing tumour extension into both posterior orbital spaces.*

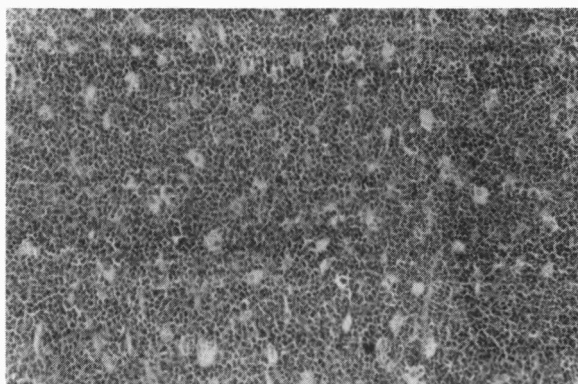


Craniotomy was performed. A large, soft tumour mass was seen in the anterior cranial fossa infiltrating the surrounding bone and brain. Removal of the tumour proved impossible, therefore only debulking was attempted.

The excised gross specimen consisted of fleshy, greyish white homogenous pieces of tissue altogether measuring 5 x 3 x 3 cms. Imprint cytology was performed and the tissue was fixed in 10 pc formalin for routine processing. Besides haematoxylin and eosin, methyl green pyronin, reticulin and periodic acid-schiff stains were done. The sections showed diffuse solid sheets of medium size blast cells with inter-

spersed macrophages giving a 'starry sky' pattern (Figure II). The blast cells corresponded with the size of the nucleus of macrophages with scanty pyronophilic cytoplasm. The nuclei were round having granular chromatic and three to five nucleoli situated mostly near the nuclear membrane. Imprint cytology revealed vacuoles in the basophilic cytoplasm. The features were consistent with the diagnosis of Burkitt's lymphoma.

*Figure II: Burkitt's lymphoma with adjacent cerebral tissue. Haematoxylin and eosin x 100.*



Improvement was seen during the immediate post operative period and the exophthalmos regressed, but deterioration in the general condition followed and the patient expired 10 days later. Permission for autopsy was refused.

## DISCUSSION

Involvement of the central nervous system is a well recognized, common complication of Burkitt's lymphoma during early relapse of tumour following treatment.<sup>1-3</sup> In relapse, the tumour infiltration is typically meningeal and subsequently parenchymal and usually presages poor outcome.

The pathogenesis of meningeal Burkitt's lymphoma relapse remains uncertain although direct extension from the primary sites has been postulated on the basis of postmortem findings.<sup>1</sup> Very rarely, an intracerebral mass can occur as a manifestation of Burkitt's lymphoma in late relapse.<sup>2</sup> The initial clinical presentation of Burkitt's lymphoma as an intracranial tumour is extremely rare<sup>7</sup> and so also is its occurrence as a primary brain tumour.<sup>4,6</sup>

Although autopsy confirmation in our patient was not done, the absence of tumour in other sites by clinical investigations was a presumptive evidence that the intracranial mass was a primary tumour.

The orbital involvement by Burkitt's tumour is usually unilateral and it can be due to a primary orbital tumour or more commonly due to invasion of the tumour from the maxillary sinus or jaw.<sup>7</sup> The ophthalmic manifestations due to the tumour include proptosis, lid changes, ptosis and cranial nerve palsies.<sup>8</sup>

Burkitt's lymphoma causing unilateral blindness has been reported by others.<sup>9</sup> The present case is unique from the point of view of the primary intracranial location and the extension into both posterior orbital spaces resulting in bilateral blindness. The damage to the optic nerves may have resulted from direct invasion by the tumour or by compression of the optic nerves from within the bony confines of posterior orbital spaces.<sup>7</sup>

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